Violaceous papules and nodules overlying bilateral dorsal hands

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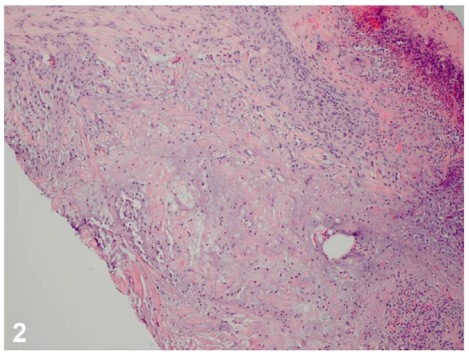

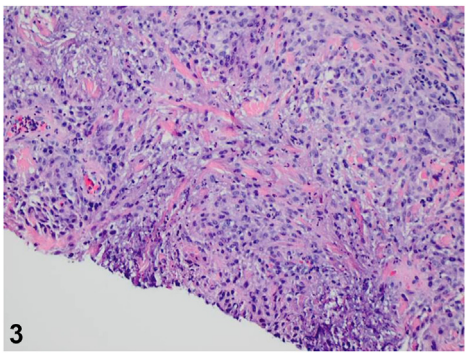

BACKGROUND

A 57-year-old man with no medical or trauma history presents with well-circumscribed violaceous papules and nodules on dorsal hands bilaterally for several years without itching, bleeding, or burning (Fig 1). Shave biopsy of the lesion demonstrated an ulcerated lesion with a neutrophilic infiltrate, some exhibiting karyorrhexis (Fig 2). There were increased spaces between the dermal collagen representing collagen degeneration with an increase in mucin deposition, histiocytes, lymphocytes, and neutrophils (Fig 3). Comprehensive metabolic panel, complete blood count, complement, sedimentation rate, rheumatoid factor,

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serum protein electrophoresis, myeloperoxidase antibodies, proteinase 3 antibodies, antinuclear antibodies, Lyme antibodies, hepatitis, and HIV testing were all negative.

Question 1: What is the diagnosis?

A. Granuloma annulare (GA)

B. Sarcoidosis

C. Interstitial granulomatous dermatitis

D. Rheumatoid nodules (RNs)

E. Palisading neutrophilic granulomatous dermatitis (PNGD)

Answer:

A. GA — Incorrect. A palisading or interstitial patchy infiltrates concentrated in the superficial and mid-dermis with focal mucin deposition would be seen in GA.[1] It is differentiated from PNGD by a relative absence of neutrophils and leukocytoclasis.[1]

B. Sarcoidosis — Incorrect. This can present with red-brown to violaceous papules and plaques most commonly on the face, however can have varying presentations. Histology shows noncaseating epithelioid granulomas with a sparse or absent surrounding lymphocytic inflammation.[1]

C. Interstitial granulomatous dermatitis — Incorrect. This presents with annular plaques or linear cords which appear on the trunk, axillae, and medial thighs in patients with rheumatoid arthritis or seronegative arthritis.[1] Histology reveals rosettes of palisading histiocytes which besiege miniscule foci of degenerated collagen with variable presence of neutrophils, absence of dermal mucin, and without evidence of vasculitis.[1]

D. RNs — Incorrect. These are firm, semi-mobile papulonodules present mostly over extensor surfaces.[1] RNs exhibit infiltrate in the deep dermis and subcutis. A palisaded layer of histiocytes and granulation tissue surrounding a central zone of eosinophilic fibrin composes RNs. An interstitial neutrophilic infiltrate can be observed in early lesions. Comparatively, neutrophils are more prominent in PNGD and are present regardless of timing.

E. PNGD — Correct. PNGD is a neutrophilic dermatosis that presents as skin-colored to erythematous umbilicated papules symmetrically distributed over the extensor surfaces.[1] The histopathological findings include small vessel vasculitis with prominent neutrophils, leukocytoclasis, or palisading granulomas.[1] There may be associated basophilic degenerated collagen.[1]

Question 2: Which of the following is not associated with the diagnosis?

A. Systemic lupus erythematosus

B. Wegener granulomatosis

C. Rheumatoid arthritis (RA)

D. Fungal infection

E. Malignancy

Answer:

A. Systemic lupus erythematosus — Incorrect. PNGD can be associated with systemic lupus erythematosus, a systemic autoimmune condition that has varying presentations from a cutaneous lesion to renal failure. PNGD has been shown to be associated with autoimmune conditions such as lupus likely due to the fact that PNGD develops as a result of perivascular immune-complex deposition triggered by systemic inflammatory pathologies.[1]

B. Wegener granulomatosis — Incorrect. PNGD can be associated with Wegener granulomatosis, a disorder characterized as granulomatous inflammation of the respiratory tracts, necrotizing small vessel vasculitis, and pauci-immune glomerulonephritis.[1] The systemic inflammatory response can trigger the development of PNGD lesions.

C. RA — Incorrect. PNGD can be associated with RA, as RA is a systemic inflammatory autoimmune condition which can cause complex deposition necessary to initiate PNGD pathology.

D. Fungal infection — Correct. PNGD is *not* associated with fungal infection. Infections such as chronic hepatitis C and Lyme disease have been reported in association with PNGD, however fungal infections specifically have not.[2,3] Other infections that have been associated with PNGD include *Streptococcus*, HIV, Epstein-Barr virus, and parvovirus.[4]

E. Malignancy — Incorrect. PNGD has been associated with lymphoproliferative malignancies such as chronic myelomonocytic leukemia, non-Hodgkin lymphoma, and Hodgkin lymphoma.[5] PNGD has been reported to develop prior to onset of these malignancies. Therefore, PNGD lesions may be an early indicator for a slew of pathologies. Because of these rare but possible associations, underlying malignancy in patients with PNGD should be ruled out, especially in those with constitutional symptoms without overt inflammatory etiology.

Question 3: Which of the following is not a treatment option?

A. Treatment of underlying disease

B. Topical steroids

C. Dapsone

D. Hydroxychloroquine

E. Ledipasvir/sofosbuvir

Answer:

A. Treatment of underlying disease — Incorrect. Treating the underlying disease has been shown to resolve the existing lesions and prevent recurrences.[1]

B. Topical steroids — Incorrect. There is no one targeted therapy for PNGD. However, there have been reports showing improvement with the use of potent corticosteroids either topically or via intralesional application.[1]

C. Dapsone — Incorrect. Dapsone has been shown to improve the lesions in a few case reports.[4]

D. Hydroxychloroquine — Incorrect. Hydroxychloroquine has also been shown to be an effective treatment for PNGD in some cases.[4]

E. Ledipasvir/sofosbuvir — Correct. Ledipasvir/sofosbuvir is *not* a treatment option for PNGD. It is an oral, direct-acting antiviral medication used to treat chronic hepatitis C virus infection which has actually been associated with the development of PNGD.[2]

Abbreviations used:

GA: granuloma annulare
PNGD: palisading neutrophilic granulomatous dermatitis
RA: rheumatoid arthritis
RN: rheumatoid nodules

Conflicts of interest

None disclosed.

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